



Neuronal Intermediate Filaments in Amyotrophic Lateral Sclerosis

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| Résumé en anglais | Neuronal intermediate filaments (NIFs) are the most abundant cytoskeletal element in mature neurons. They are composed of different protein subunits encoded by separate genes such as neurofilament light chain (NFL), neurofilament medium chain (NFM), neurofilament heavy chain (NFH), α -internexin and peripherin. NIFs are dynamic structures playing important functions in cell architecture and differentiation, interactions between proteins or subcellular organelles, and in axonal calibre determination and myelination. Consequently, their presence modulates electrophysiological properties of axons. NIFs have long been assigned a role in the pathogenesis of amyotrophic lateral sclerosis (ALS). Indeed, accumulation and abnormal phosphorylation of NIF subunits in motor neuron are one of the major pathological features in both sporadic and familial forms of the disease. Moreover, mutations in the NFH and peripherin genes and elevated cerebrospinal fluid NIF levels reported in ALS cases, associated with studies in transgenic mice, provided the evidence that primary defects in NIFs could be causative for motor neuron disease. However, the processes leading to the NIF abnormalities and the links to the pathogenesis of ALS remain unclear, leaving a challenging open field for further investigations in this highly disabling disease. Here, we review the main characteristics of these NIFs and their involvement in the pathomechanisms of ALS. |
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- [1] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=26274>
- [2] <http://okina.univ-angers.fr/julien.cassereau/publications>
- [3] <http://okina.univ-angers.fr/joel.eyer/publications>
- [4] <http://okina.univ-angers.fr/franck.letournel/publications>
- [5] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=26276>
- [6] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=26277>
- [7] <http://okina.univ-angers.fr/publications?f%5Bkeyword%5D=22366>
- [8] <http://okina.univ-angers.fr/publications?f%5Bkeyword%5D=22368>
- [9] <http://okina.univ-angers.fr/publications?f%5Bkeyword%5D=22365>
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- [12] <http://okina.univ-angers.fr/publications?f%5Bkeyword%5D=14644>
- [13] <http://okina.univ-angers.fr/publications?f%5Bkeyword%5D=14259>
- [14] <http://okina.univ-angers.fr/publications/ua15633>
- [15] <http://dx.doi.org/10.5772/63161>
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